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CONTINUING MEDICAL EDUCATION
ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 46

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2016, 33(2):282–284

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A 68-year-old woman was admitted to the hospital because of 6 month history of bone pain located in lumbar area and aggravated by movement, as well as fatigue. The non-steroidal anti-inflammatory drugs (NSAIDs) administration had no effect. The patient also presented progressive weakness and mild dyspnea on exertion. She, occasionally, was feeling dizzy and experienced visual disturbances. One week before admission she had an episode of nosebleeding. Her past medical history was unremarkable.

On examination, she appeared pale. The temperature was 38.8 °C, the pulse was 110/min and the blood pressure was 11/70 mmHg. The liver was non-tender, and palpable 2 cm below the right costal margin. The spleen was not palpable. Pressure on the lumbosacral vertebrae worsened the pain, which had radicular features. Her hematological tests revealed a normocytic normochromic anemia (Ht: 26.9%, Hb: 8.5 g/dL, with 0.1% reticulocytes, a mild leukopenia (WBC 3.100/μL, neutrophils 19%, lymphocytes 42%, monocytes 9%, metamyelocytes 4%, and atypical blast cells 24% (figures 1 to 5) and platelets 40.000/mL. The erythrocyte sedimentation rate was 125 mm/1 hour. Coagulation studies were normal. Her biochemical tests were: BUN 75 mg/dL, creatinine 2.6 mg/dL, SGOT 19 IU/L, SGPT 21 IU/L, LDH 1,110 IU/L, ALP 135 IU/L, γ-GT 29 IU/L, Na^+ 141 Meq/L, K^+ 4.7 Meq/L, Ca^{++} 6.3 mg/dL, and serum total proteins 10.9 g/dL (albumin 4 g/dL, globulins 6.9 g/dL). The serum protein electrophoresis revealed a spike in the area of γ-globulins. The bone marrow aspirate was diagnostic (fig. 6). On the second day of hospitalization, the patient's condition worsened. The temperature rose up to 39.4 °C, and she presented a mild pain in the right arm and thigh

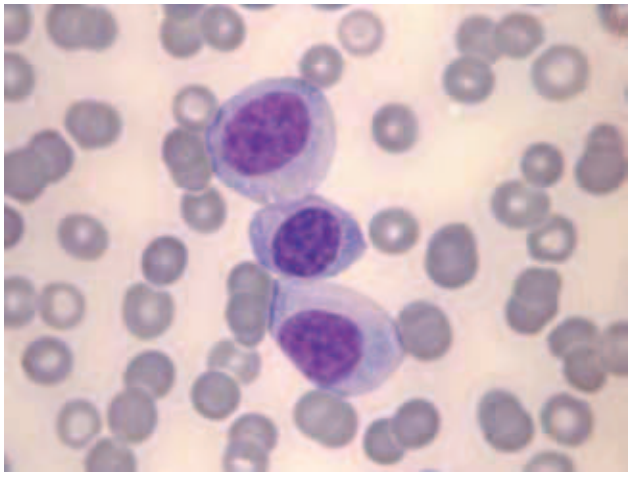

Figure 1

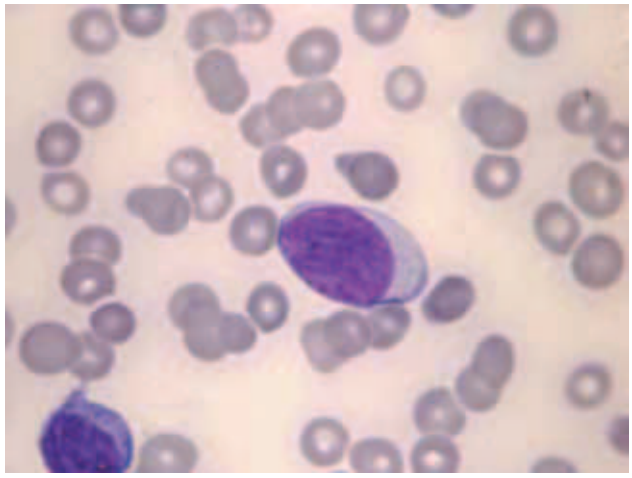

Figure 2

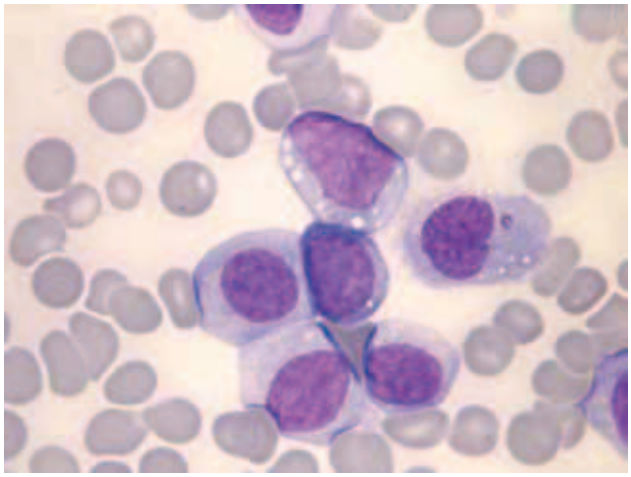

Figure 3

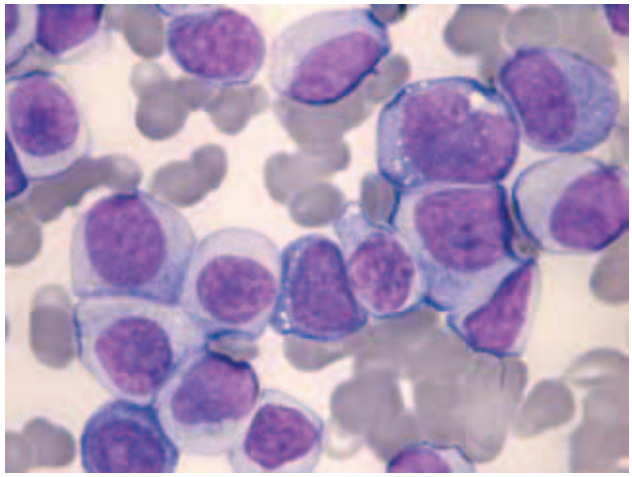

Figure 6

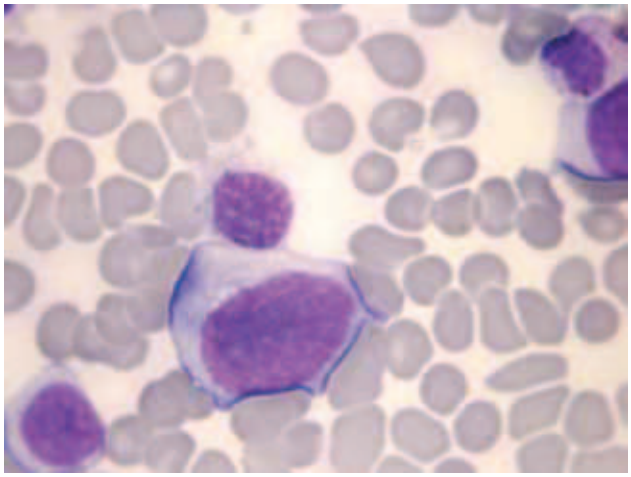

Figure 4

with contemporary presence of redness and warm. Rapidly the patient's condition deteriorated further, as rigors were added and she became disoriented. The pulse was very weak and the blood pressure was 80/40 mmHg. Antibiotic therapy with a combination of ceftazidime plus amikacin was started. The skin lesions on the arm and on the other body areas enlarged and gradually small ulcers containing a greenish smear were formed (figures 7 to 9). The patient's condition improved after 4 days on antibiotic treatment. Blood, as well lesion cultures were positive.

Comment

Plasma cell leukemia is a rare and often aggressive disease because of clonal proliferation of abnormal plasma cells with phenotypical abnormalities similar to those present in multiple myeloma. The

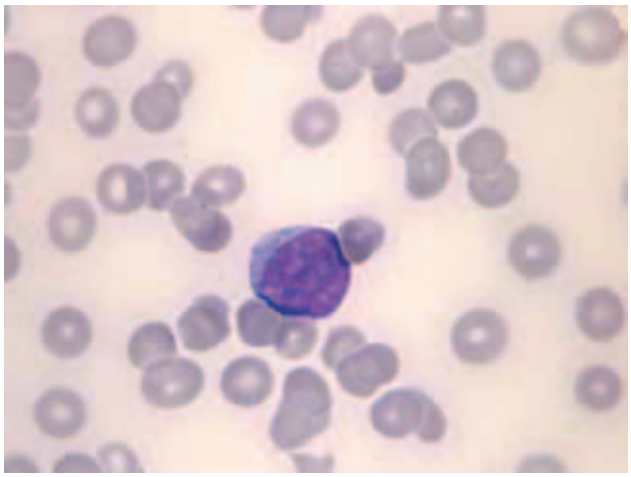

Figure 5

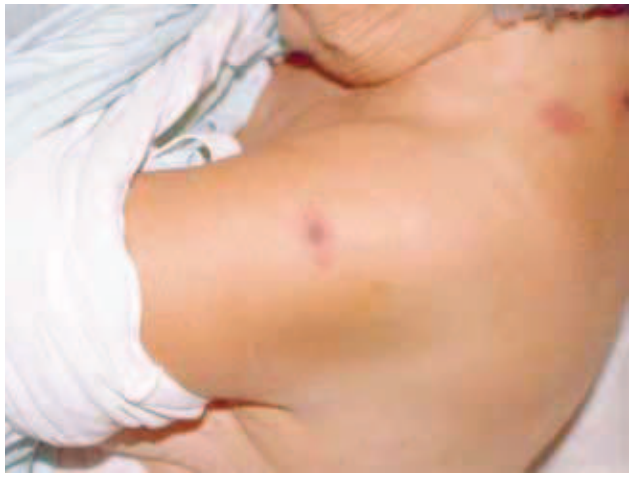

Figure 7

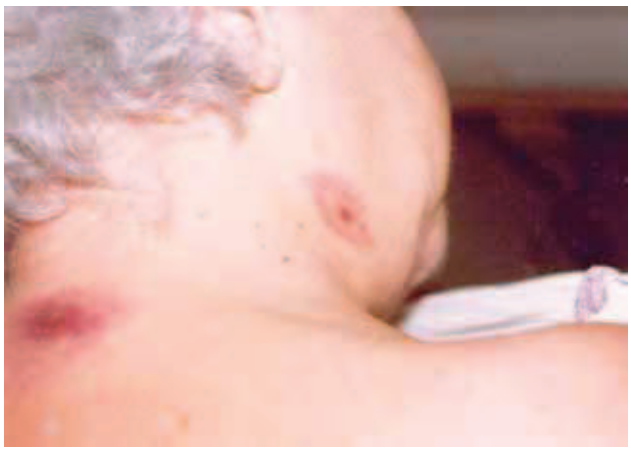

Figure 8

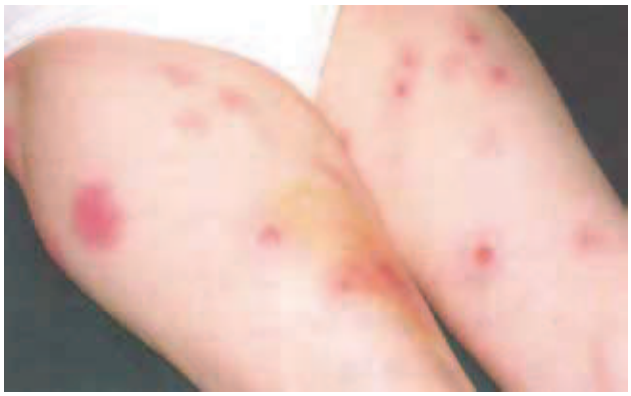

Figure 9

condition is characterized by the presence of numerous plasmacytes and or plasmablasts in the peripheral blood (>2.000/μL, at least 20% of cells). In some patients, the circulating cells have a small size varying from the mature lymphocytes with basophilic cytoplasm to plasmacytes with abnormal appearance and or plasmablasts, and accompanied anemia and thrombocytopenia. This picture is present in extramedullary plasmacytomas with concomitant hepatospleenomegaly. At times the cells are difficult to differentiate from an undifferentiated acute leukemia of a leukemic phase of large cell non-Hodgkin's lymphoma (distinction by the cytoplasmic expression of cytoplasmic immunoglobulin, absence of normal B cell antigens and expression of the late B-cell antigens such as the CD38). Plasma cells are phenotypically CD138+/CD38+/CD45– or weakly positive, while CD56 is frequently negative, CD117 and HLADR are also less frequently expressed in plasma cell leukemia and CD20 is more frequently present. In cases of circulating large numbers of small leukemic plasmacytes, their distinction from the large lymphocytes or prolymphocytes, by the existence of a well visible perinuclear halo and the characteristic basophilia, especially in the peripheral cell cytoplasmic area of plasmacytes is necessary. The bone marrow is infiltrated by similar blastic cells. Plasma cell leukemia may be present de novo or as evolution of a multiple myeloma.

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